A MICROSCOPICAL STUDY OF THE SPINAL CORD, NOT COMPRESSED BY DISPLACED VERTEBRÆ, IN A CASE OF POTT'S DISEASE; AREAS OF NECROSIS IN THE CEREBELLUM, ONE SUPERIOR CEREBELLAR PEDUNCLE AND CORD; RENAUT'S BODIES IN ONE SCIATIC NERVE.

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The case I am about to report is interesting from several standpoints. Besides the uncommon pathological findings we have here a variety of Pott's disease, in which the cord was entirely free in the spinal canal and no distinct adhesions of the meninges were noticeable. From the absence of compression of the cord one could infer that it is a case of Pott's disease in its earlier stages, but the changes found point rather to a disease of long duration. The latter tend to show that toxins may affect the cord to a considerable extent long before vertebral or pachymeningeal compression takes place. This observation particularly deserves our attention.

J. H. C—, aged fifty-three, colored, steward in U. S. Navy, admitted to the Douglass Memorial Hospital, January 19, 1903. He gave the following history:

Fifteen years ago had a chancre and orchitis. Was married twice, both wives had miscarriages. In August, 1902, while at work on the ship, he was exposed for a long time to dampness, after which he suffered intense pain in the lumbar region. Three weeks of treatment in the German Hospital improved the condition, but he soon developed severe pain in the epigastrium down to the umbilicus. Shortly afterwards the pain extended to the lower extremities and since then the latter gradualy became numb and weak.

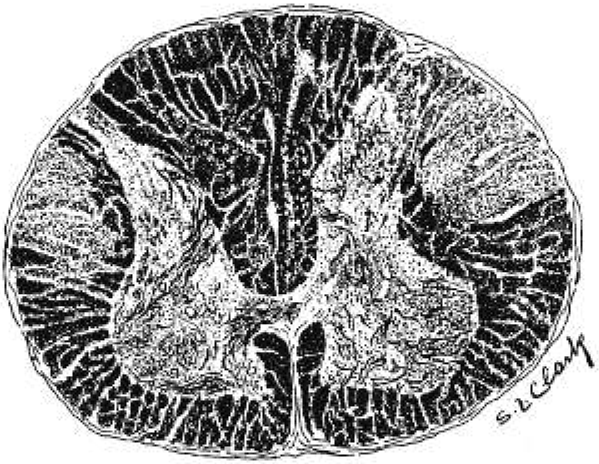
The examination on admission showed complete flaccid paraplegia with the knee-jerk lost on the left side, exaggerated on the right, no ankle-clonus on either side, Babinski only on the left side. Sensory disturbances were marked: touch and pain sense abolished on the right, delayed on the left, temperature sense lost on both sides. Patient presented marked trophic disturbances of the skin: two bedsores of a few weeks' standing and the nails were very much deformed and brittle. The sensations of the perineum were normal. The abdomen below the umbilicus presented complete loss of all sensations with the exception of a few areas which were hypesthetic. Above the umbilicus on the trunk and upper extremities were no sensory or motor disturbances. The sphincters of the bladder and rectum were much involved: very frequent and imperative micturition, also incontinence of feces disturbed the patient considerably. The pain of the lower extremities mentioned above disappeared entirely. As the patient complained of a dull pain in the back between the shoulders, the spinal column was examined and a prominence of the 9th and 10th dorsal vertebræ was found. The upper extremities showed distinct muscular atrophy, especially in the small muscles of the hands.

A second examination of the patient made a fortnight later showed that the knee-jerk on the right, which was before exaggerated, was now very much diminished. Patient also complained of tingling sensations, and at times of pain in the left upper extremity. The muscular atrophy of the upper limbs was still more marked.

An examination made some time later showed total disappearance of the right knee-jerk and increased sharp pain in the left arm. The muscular atrophy was extreme, so that he handled objects with great difficulty. The bedsores suppurated considerably. The patient began to cough and expectorate and had occasionally evening chills. There was slight elevations of the temperature in the evening (99.3°—100°). Sputum showed tubercle bacilli. The pain in the back gradually increased, the condition of the sphincters became intolerable, and on the 14th of April a right facial paralysis with a marked weakness of the right arm and inability of swallowing made their appearance. The patient soon became unconscious and expired. The autopsy was made 12 hours later by Dr. A. G. Ellis, and these are the gross findings he reports:

AUTOPSY FINDINGS ON BODY OF MR. C—, DOUGLASS MEMORIAL HOSPITAL, APRIL 14, 1903.

"The body is that of a markedly emaciated adult colored male. There is a large irregular ulcer over the sacral region with extensions well down over each buttock. There is a noticeable, but not marked, prominence of the spines of the ninth, tenth and eleventh dorsal vertebræ. On opening the spinal canal there is found caries and softening of the bodies of these vertebræ, especially the ninth and tenth. Angulation is not present and the lumen of the canal, although slightly encroached upon by the carious vertebræ, is not sufficiently interfered with to cause noticeable compression of the cord.

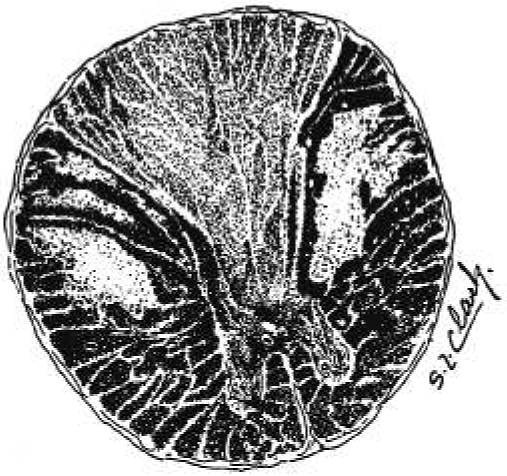


Lumbar segment showing descending sensory degeneration in the posterior columns, also degeneration of crossed pyramidal tracts.

The lower portions of the cord are perhaps slightly swollen. The superior surface of the brain is somewhat edematous, but shows no conspicuous gross lesion.

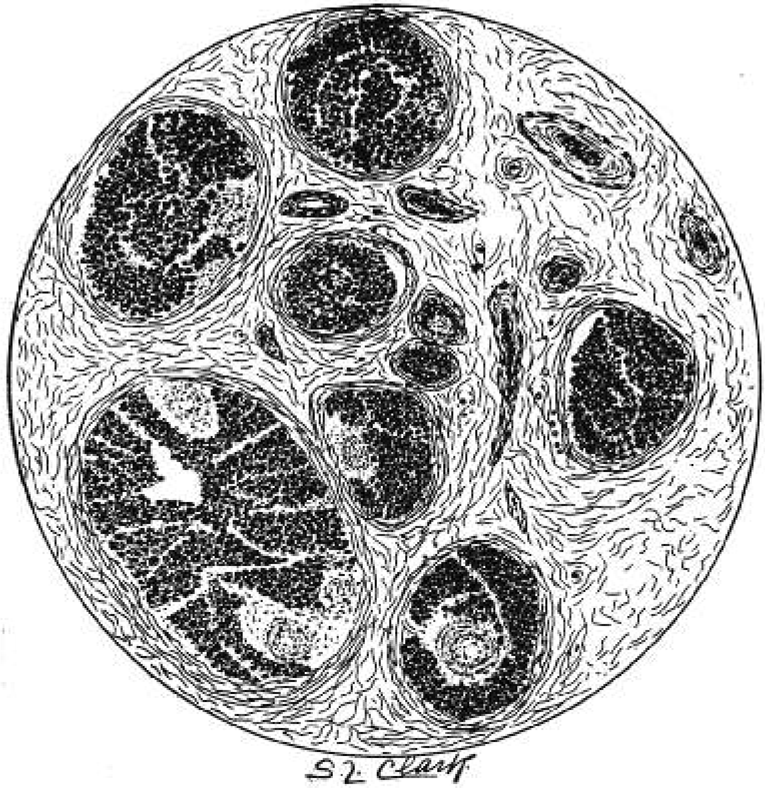
"The lungs show some emphysema, with hypostatic congestion of the posterior portions. The middle lobe of the right lung is adherent to a protuberance on the vertebral column at a point corresponding to the level of the carious vertebræ mentioned. On separating these adhesions the lung is torn, revealing a softened, caseous area 5 cm. in diameter. The surrounding lung tissue contains numerous gray nodules, none of which are more than 0.5 cm. in diameter. The left side of the vertebral column at this point shows two small protuberances, but the lung is not adherent and is apparently sound. The kidneys show changes indicative of a moderate degree of chronic diffuse nephritis. The liver is congested and shows evidence of fatty infiltration. The other viscera show no gross lesions."

The brain, the cord (except its cervical portion, which for some reason was not removed), a portion of the lung, sections of both median and sciatic nerves were kept for microscopical studies. The lung showed caseous areas encapsuled in connective tissue. Sections of the brain showed nothing of importance. The cerebellum on horizontal sections showed in the center a large area of softening. This was more marked on the right half than on the left and included also the middle lobe. The softening extended down to the lower surface of the cerebellum, so that it can be safely said that a part of the roof of the 4th ventricle, was formed by the pulpy mass of the softened cerebellum. The softening occupied only three-fourths of the thickness of the cerebellum. After the usual process of hardening, the tissues were examined microscopically with the methods of Marchi, Weigert, Weigert-Pal, thionin, hemalum and carmin.



Tenth thoracic segment, showing areas of necrosis in the lateral tracts, also complete degeneration of posterior columns.

The cells of the entire cord showed very pronounced changes, and particularly in the mid-thoracic region. In some cells the periphery stains less than the center, they have lost their multipolar appearance, are oval in shape. Some cells present very fine granulations (dust-like), one pole is discolored, in some the chromatic substance is broken up and not uniformly distributed through the cell, no dendrites, nucleus excentric, nucleolus absent. Other cells present a central chromatolysis with a nucleus irregular in shape and position, without a nucleolus. Some cells are very much diminished in size (atrophy) with chromatin only around the nucleus, which forms hernia on the periphery of the cell. Other cells are very much elongated with chromatin only on the poles, vacuoles on one side, dendrites hypertrophied, two nuclei. On some sections only vestiges of cells can be seen, or round cells with torn off edges, empty in the center, free from nuclei. In the upper and mid-thoracic sections are found quite a number of cells filled with yellow pigment (pigment degeneration) without dendrites or nuclei, with marked deformities. In the upper lumbar sections there is also a marked



Right sciatic n, showing Rénaut's bodies.

chromatolysis with deformities of the cells, but these changes are less marked than in the sections above. In Clarke's columns there are very few cells at the level of the 10th and 11th thoracic segments. In sections below this level distinct chromatolysis could be seen.

The spinal ganglia are markedly diseased. Three-fourths of the cells are empty (no granules), nucleus and nucleolus lie either on the periphery of the cell or are entirely absent. All the sections of the entire cord show the presence of a great number of amyloid bodies, especially in the posterior columns.

Examination for Degeneration:—Upper thoracic cord.—The degeneration, is distinctly limited to Goll's tracts except posteriorly, where it encroaches slightly upon Burdach's columns. It is also present in the posterior roots as they approach the tips of the posterior cornua. Near the roots there are dilated and obstructed blood vessels. Some degenerative changes are noticed in the anterior roots with similar changes in blood vessels. Near the central canal two cavities are seen. With Marchi this section shows some degenerative changes also in Burdach's columns, and by far more degenerated nerve bundles than with Weigert's method.

Ninth thoracic segment.—The degenerated area in the posterior columns has increased considerably and involved also the entire Burdach's tract, except in its postero-lateral portion. The direct cerebellar tract in its anterior portion also that portion of Gowers' tract with which the latter is in contact, show some degeneration. The posterior roots are not involved in this section, but the anterior show the same changes as in the previous section. With Marchi Gowers' tract is seen to be more involved than with Weigert's method. In Lissauer's tract also all the nerve bundles show considerable degeneration.

Tenth thoracic segment shows complete degeneration of the posterior tracts, two small cavities near the central canal, foci of softening in the lateral columns corresponding strictly to the position of the crossed pyramidal tracts. Posterior roots show distinct degeneration on one side of the cord. Here again numerous dilated and thrombotic blood vessels in the nerve bundles and at the periphery of the cord.

Eleventh thoracic segment.—In the posterior columns besides a few degenerated fibers there are seven or eight foci of softening on one side and three or four on the other disseminated in the columns of Goll and Burdach. Two areas of softening we find in the anterior columns corresponding to the position of the tracts of Löwenthal and Marchi and the olivary tract. In the lateral columns there is some degeneration of the cross pyramidal tract involving also partly the very posterior portion of Gowers' tract. The posterior roots are somewhat degenerated. At the periphery of the cord are seen dilated blood vessels, the majority of which are filled with thrombi; some nerve bundles around the cord are degenerated and contain dilated and thrombotic blood vessels. With Marchi the direct cerebellar and Lissauer's tracts show considerable degeneration.

First lumbar segment presents small cavities near the central canal, a linear degeneration in the posterior columns of one-half of the cord, but distinct degeneration of the crossed pyramidal tracts and only moderate changes in the direct cerebellar tracts. The posterior roots show no degeneration. The condition of the blood vessels is the same as in the previous sections. The linear degenerated area begins below the commissure and is somewhat diffuse at this level, but when it reaches the middle of the posterior columns, it becomes distinctly linear and is located almost close to the septum; at the lower third of the latter it moves over towards the middle of the column where it becomes again diffuse without, however, reaching the periphery of the cord.

The second lumbar presents also cavities and the same linear degeneration of the posterior columns, although smaller in size. The direct cerebellar tract does not show any involvement.

In the fifth lumbar segment the linear degeneration is no more in its former place, and in its place we find a similar long area midway between the septum and the cornua, although nearer the latter than in the 1st, shorter and wider than in the 1st and 2nd lumbar segments. Again the commissure and the periphery of the cord are not touched. On the same section we also notice some diffuse but not marked alterations near the commissure on either side of the septum. The roots are not involved.

As to the meninges, changes were found mainly in the 10th and 11th thoracic segments, but also to some extent in the lumbar portion. They consist of a distinct, although not very pronounced thickening and leucocytic infiltration of the dura, and to some extent of the pia. The blood vessels at the periphery of the cord, besides their very marked dilatation, show distinct endoperiarteritis with small cell infiltration. The latter was also found in the perineurium of the nerve roots in some sections. The small cell infiltration can also be seen in the vessels of the degenerated areas in the cord, especially of the 10th and 11th segments.

In the medulla no changes were found in the nervous tissue, but the blood vessels were distinctly dilated and some of them filled with thrombi. In the upper portion of the pons two areas of softening are seen in one of the superior peduncles.

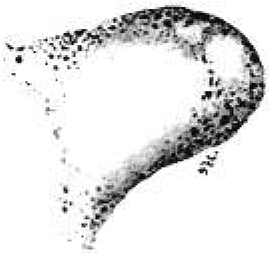
The examination of the peripheral nerves show that both median and the left sciatic nerves present some degenerative changes, thickened epineurium and in some places enlarged blood vessels. In examining the right sciatic nerve I found structures in some of the nerve bundles which resemble those described by Rénaut in 1881 and by Spiller in 1900. Curiously enough no degenerated areas could be traced in this nerve.

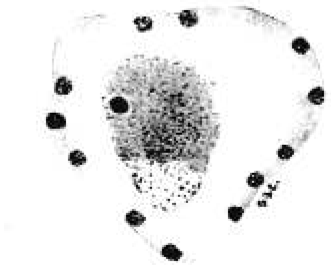
The degenerative changes described in the cord appear to have their point of departure in the 10th and 11th thoracic segments. As we have seen, the foci of softening in the 10th segment lay in the lateral columns in the region of the pyramidal tract. Below this level in the 11th thoracic we see already some typical degenerated fibers in the crossed pyramidal tract and in the lumbar segments complete descending degeneration of this tract. Above this segment the crossed pyramidal tract is intact. In the 11th thoracic segment the foci of necrosis are concentrated in the posterior columns with the result, that in sections above this level the posterior columns show complete degeneration. Up to this point the pathological changes are found to be in accordance with our classical knowledge of the direction of the sensory and motor tracts. Below the 11th thoracic segment, however, I found a linear degenerated area in the posterior columns, corresponding in some respects to the description given by Schultze (*Arch. f. Psych. u. Nervenkr.,* 1883). This writer in studying a transverse lesion of the cord found below the diseased focus in the posterior columns a degenerated area which he described as two lines parallel with the posterior cornua from which they lie at a certain distance and do not reach the periphery of the cord. In my sections from the 12th dorsal down to the sacral segments this linear stainless area can be seen distinctly with Weigert's method.

According to some authors this so-called comma area of Schultze reaches the periphery of the cord and the posterior commissure, some on the other hand say that it occupies only one portion of Goll's columns. Dejerine has shown that in degeneration of the roots Schultze's bundle is smaller in length and width than in cases of transverse myelitis. Various descriptions of isolated degenerated tracts in the posterior columns have been given under the same name of comma zone of Schultze, although they do not all correspond to the exact position as given by Schultze. At all events the occurrence of an isolated linear degenerated area in the posterior tracts recalls the one described originally by Schultze. The significance of it is that it is the expression of a descending sensory degeneration. Although Schultze's case was the first to show the comma-like degeneration bearing his name, nevertheless descending degenerations in Goll's columns were known long before him. Bastian (*Med.-Chir. Trans.*, 1867), Kahler, Pick (*Arch. f. Psych.*, 1880), Strümpell (ibid.) and Westphal made this observation before Schultze, and at present this is a fact accepted by all neuropathologists. Before Hoche's publication in 1896 (*Arch. f. Psych.*) this descending degeneration of Schultze's type was not observed below the 11th and 12th thoracic segments. This author and after him Senator (*Ztschr. f. Klin. Med.*, 1898) who corroborated his findings have shown that in lesions as high as the 7th thoracic (Hoche) and even as the 7th cervical (Senator), this comma like descending degeneration may begin as low as the 10th thoracic (Hoche) and continue down to the lower lumbar segments. Dejerine and Théohari, after a careful analysis of all the cases reported by others and of their own arrive at this conclusion, that the comma-like tract can be found in any portion of the cord; its localization depends upon the situation of the initial lesion; in lower transverse lesions the area in question will be found even as low as the sacral segments. In my case the descending degenerated area corresponds in many points to the classical comma-type of Schultze. As to the explanation of this retrograde degeneration we must refer mainly to the works of Cajal, v. Lenhossék and Kölliker, who have shown that the posterior root fibers in entering the cord are divided into ascending (long) and descending (short) branches. In root lesions therefore we can expect degenerations of these two branches and especially of the short ones. The latter condition probably will explain the cause of the existence of Schultze's comma zone. However, not all neuropathologists are agreed on the origin of the long and short fibers in the posterior tracts. In my case, for example, the sections containing the descending degenerated area did not show root involvement. The direct cerebellar tract was also involved to a certain extent below the 11th thoracic segment, which can be better seen with the Marchi stain. However, this does not extend below the 3rd lumbar segment. We have here, therefore, again a descending sensory degeneration.

The case presents another interesting feature. When in Pott's disease the angle formed by the displaced vertebræ reaches the meninges and the cord, the pathological changes of the latter may be argued as due to direct pressure, although this point is not entirely settled. Cases of this character are abundant in the literature. But cases of Pott's disease in which the changes of the cord are marked in spite of absence of all trace of compression, are comparatively rare. In my case the cord was entirely free in the spinal canal and still the pathological changes are very pronounced. It raises at once the question of the possibility of influence at a distance of the tuberculous process of the bones. Moreover, it also shows that pronounced cord changes may be present with only slight meningeal involvement, as the meninges as stated above are moderately affected even at the level of the highest changes in the cord.

Let us analyze first of all the nature of the pathological findings. The thickening of the meninges and their leucocytic infiltration as shown with a nuclear stain (hemalum) especially in the walls of the blood vessels presents nothing specific. The condition as it is seen is found in tuberculosis as well as in syphilis. In the degenerated areas of the cord itself we also find small cell infiltration of some blood vessels, besides the areas of necrosis described above. The striking feature of the histological changes concerns the blood vessels, which are equally affected in the meninges, cord and nerve bundles surrounding the cord. Extreme dilatation in some places, considerable dilatation in most of the sections, thickening of the adventitia in the majority, disappearance of the intima in some, thrombi in the majority of the vessels, hemorrhagic foci around some arterioles, besides the leucocytic infiltration mentioned above—these are the prominent changes found not only in the cord with its meninges, but also in the medulla, pons and cerebellum. Such a condition of blood vessels suggests at once tuberculosis of the nervous system. While on one hand no typical tubercles with giant cells were found, also repeated attempts at staining the nervous tissue for tubercle bacilli failed, on the other hand the necrotic areas in the cord situated at the same level with the caseous vertebræ, the condition of the arteries and veins which has been found in the majority if not in all cases of tuberculosis of the cord, in addition to the fact that the patient's lung was distinctly tuberculous and the sputum showed tubercle bacilli, —all these facts are in favor of tuberculosis of the nervous system in our case. There are now on record cases showing beyond doubt that the cord may be affected with tuberculosis without the presence of giant cells or miliary tubercles or even tubercle bacilli. Histologically our case resembles in many respects the one of the three published by Philippe and Cestan, who proved the tuberculous nature of the cord by experimental inoculations. In their 3rd case they have found total absence of pachymeningitis and still disseminated foci of parenchymatous myelitis were present. A careful analysis of all the microscopical records concerning Pott's disease published since show that the pathological process may vary considerably: myelitis in Potts' disease may be due to compression from the vertebræ, from peripachymeningitis, may be due to a moderate meningitis or even the cord may become primarily involved without participation of the meninges under the influence of tuberculous toxins circulating in the blood vessels of the cord. Here I wish to call attention to a very suggestive histological point noticed in my case. The endovascular changes are far less marked than the perivascular, a fact which is probably in favor of a perivascular lymphatic origin of the tuberculous process. Homen's recent experiments (*Neurol. Centralbl.*, No. 3, 1904, p. 21) prove conclusively the lymphatic route of injected bacilli and of their toxins; they also show that while in recent cases the pathological changes





of the nervous system depend upon the localization of the bacteria, in older cases in which the bacteria can no more be found, the alterations depend upon the poisons elaborated by the bacteria.

In considering the character of the findings we must not forget that syphilis may in a great measure simulate tuberculosis of the meninges and cord. Indeed, from histological standpoint syphilitic and tuberculous meningomyelitis do not differ from each other at the beginning. The endoperiarteritis and phlebitis, small cell infiltrations may be encountered in both affections. At this period it is only the etiology which enables us to make a differentiation. However, the tendency to ruptures of blood vessels and hemorrhages are more in favor of syphilis. In my case the diffuse myelitis was very marked, consequently at this period of development we should expect distinct gummatous formation, unless again like in tuberculosis of the cord the characteristic pathologic formation may be absent and the case still be one of syphilis. At all events my patient presented a suggestive history of syphilis and perhaps the changes in the blood vessels are the expression of a mixed infection.

As a last peculiarity noticed in the pathological findings of the present case is the so-called Rénaut's bodies. According to Spiller (*J. of Exper. Med.* vol. v., No. 1) Rénaut was not the first to mention these peculiar structures, which, however, bear the name of the writer who studied them in particular. They are found in nerves and present on transverse sections concentric circles of connective tissue with nuclei which are also concentrically arranged. I found these bodies only in the right sciatic nerve, in which there was no trace of ordinary degeneration. Both median and the left sciatic nerves which were also examined did not contain Rénaut's bodies. Spiller, who to my knowledge was the first in this country to describe these structures, found them only in one of the nerves of the brachial plexus. The latter seems to be a favorite place for them. As to the function of Rénaut's bodies, there is a theory according to which they serve as a protection for the nerve fibers, but this view is not shared by Spiller and others who have studied them in detail. It is only known that they are found in normal and pathological nerves. In my case the nerve which contains these bodies presents no typical degenerative areas. Prof. Coplin, who has seen these bodies in sections from my case, expressed an opinion that they resemble somewhat Pacinian corpuscles, which are sometimes found on the nerves.

As a last point of interest is perhaps the condition of the reflexes in the lower extremities. At first alongside of complete flaccid paralysis Babinski's reflex was present on the left side, where the knee-jerk was lost, but not on the side where the knee-jerk was exaggerated. Later all these exalted reflex-phenomena disappeared. The present case is not suitable for any deductions on the pathogenesis of reflexes. We know now that the reflex mechanism has multiple relations and the analysis of the accumulated observations (experimental and clinical) tends more and more to show that the exclusivism of Bastian, Crocq, Collier and Buzzard cannot be accepted, but that Grasset's views are more in conformity with facts, namely, that spinal, basilar and cortical regions contain centers for both reflexes and tonicity; the latter undoubtedly has some controlling influence upon the former. The total loss of all the reflexes in our case has perhaps some interrelation as cause and effect with the softening of the cerebellum and one of the superior peduncles in addition to the diffuse myelitis.

EXPLANATION OF PLATES

From above downward—Cell of lumbar segment. Cell of spinal glanglion. Cell of lower thoracic segment.